

Lidell J. A.

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OF

NEUROMA OF THE OPTIC NERVE,

WITH REMARKS AND ILLUSTRATIONS.

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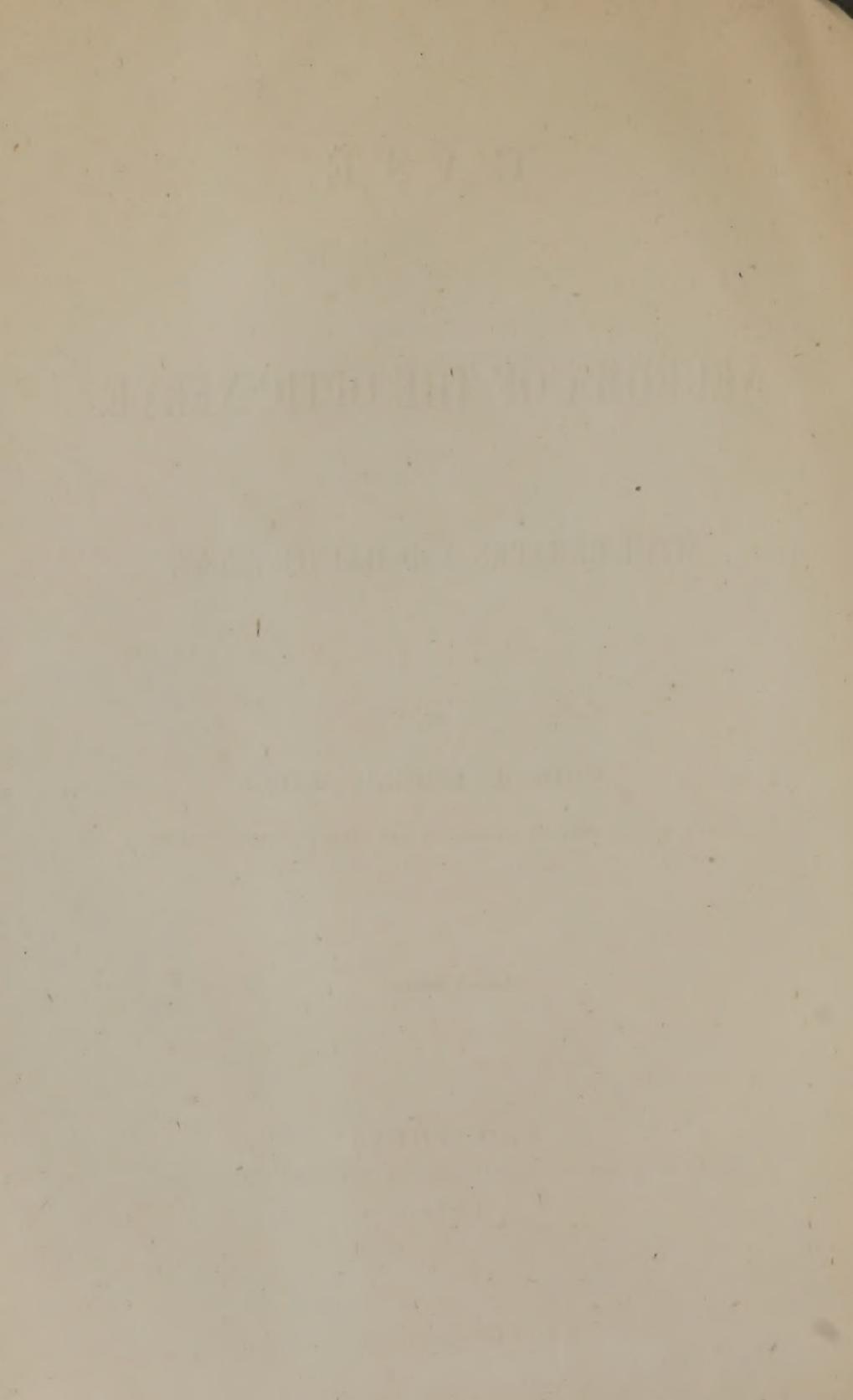
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A CASE
OF
NEUROMA OF THE OPTIC NERVE.

Neuroma (from *νεῦρον*, a nerve) signifies a benign tumor developed in connection with a nerve. This disease is not often met with ; and it is only within a very recent period that the general character and relations of these growths have been commonly understood, and that they have been arranged together in a special group or class, denominated neuroma. By the older surgeons, all tumors of nerves were considered to partake of a malignant character, and even Chelius, in his great work on surgery, although he distinctly recognizes the non-malignancy of the nerve-tumors under consideration, still describes them under the head of, and in connection with, cancer of the nerves. The excellent monograph of Mr. Robert W. Smith, of Dublin, gives the fullest, most recent, and best account of neuroma which we have seen, and, as Mr. Erichsen well remarks, it contains "the principal part of our knowledge of this subject."

Neuromata are not confined to any one part of the body, to the exclusion of all other parts. They are, however, more frequently met with in the extremities than in the trunk ; and, again, found oftener in the lower than in the upper extremity. Their principal seat is the *cerebro-spinal* system of nerves ; though Bérard has once met with this morbid growth on a *ganglionic* nerve. The case about to be described is the *only one* I have been able to find where this disease involved the *optic nerve*, and almost the only one involving a nerve of special sense.

ture. 2. It never contaminates the neighboring lymphatic ganglia ; carcinoma does. 3. While it impairs the patient's general health by pain and distress, causing pallor, emaciation, and debility, it never produces the true cancerous cachexy by specific infection of the system ; carcinoma does produce such a result. 4. It does not return after excision ; carcinoma does. 5. The pain of neuroma is neuralgic (electrical and paroxysmal) in character ; the pain of carcinoma is not.*

The comparative rarity of the neuromatous disease, its entire benignity, the liability to mistake a bad case of it in an advanced stage for malignant disease, the fact that it is not amenable to *medical* treatment, that it can not be cured short of extirpation, and that it is cured by operation with great certainty, according to the testimony of all observers, and, finally, the fact that I have not been able to find the history or the description of any other case of neuroma, involving the optic nerve, have induced me to place the following case on record, as a contribution to the current surgical literature of the day. It is barely possible that individual cases of neuroma of the optic nerve have passed hitherto for cancer of the eye, or orbit, or both, to which, indeed, in an advanced stage, it bears some resemblance, and thus have either not been extirpated, or, if extirpation has been practiced, the disease has borne another name.

Case.—In the latter part of July, 1858, a pale and rather spare young woman, of twenty years, named J. L., applied to me for advice concerning a large, red, fleshy-looking, elongated, and rounded mass, which filled the left orbit, and protruded therefrom, so as to hang over the cheek almost as far as the level of the nostril [vide plate].

She and her mother together gave the following history : Her health had been good till the spring of 1850, when she was twelve years old. Previously, she had had measles, scarlet fever, and hooping-cough, but had them so light, as not to produce any difficulty worth mentioning. In the spring of 1850, she, along with other children, took the mumps (parotitis), which involved the gland on both sides of the neck. The difficulty in the right gland followed the usual course, which was not the case, however, with the one on the left side. The swelling in this gland did not readily subside, but it remained enlarged and troublesome for nearly a year. In the latter part of the same summer (1850) the left eyelids became swelled. At the same time the right eyelids were natural. This palpebral tumefaction lasted two or three months, or till

* Vide Smith's monograph.

the last of autumn. At first her mother supposed it was produced by poisoning with ivy (*rhus radicans*—they lived in the country), but this conjecture was negatived by the fact that the swelling was confined to the left eyelids, and lasted for so long a time. Four or five months after the palpebral swelling had disappeared, that is, in the spring of 1851, she found the sight of the left eye was becoming dim. Objects, when seen through this eye alone, appeared to be in a mist, or haze. She could, however, distinguish the scholars, as they sat in their places in school, so as to recognize them. At the same time the sight in the right eye was unimpaired. The left parotid gland still continued to be enlarged. Through the remainder of spring, and the following summer, the dimness of sight (amblyopia) increased so rapidly that in the fall (1858) she could only distinguish light from darkness. During all this time she did not suffer any pain, nor *muscae volitantes*, nor colored spots, nor flashes of light in the affected eye, so far as she remembers. About this period, *i.e.*, some time in the fall of 1851, she noticed that this same left eye began to protrude (exophthalmos). After this, the perception of light rapidly became fainter, and soon disappeared; the eye gradually protruded more, until, twenty months afterward (June, 1853), the eye protruded so much that the lid could not be closed over it. Her mother says that, at this time, the eyeball itself did not seem to be much enlarged; also, that the pupil was widely dilated, and that every thing behind the pupil appeared to be dark-colored. She was positive she never saw any yellowish or shining metallic appearance behind the pupil. The patient says that at this time the eye was distinctly movable in front of and upon the substance, whatever its nature might be, which caused the protrusion, and had been thus movable for some time previously. In this statement she was very positive. Up to this time she had not suffered any pain, except about a year before, when a compress was applied by the late Dr. White, of Cherry Valley. Having tried various remedies without benefit, he resorted to compression, for the purpose of arresting the protrusion, but, after continuing it about two weeks, was forced to abandon it, on account of the severe pain it occasioned. After the compress was removed, the pain ceased. Up to this time her general health had not suffered.

In the latter part of June, 1853, her symptoms underwent a material change. On the day subsequent to a long walk (five miles) under a hot and bright sun, she had a good deal of pain in the affected eye, which became red, and speedily swelled to a great size (conjunctivitis). The eyelids also were much swelled, and purplish in color. During the following fortnight

she suffered very much from the severity of the inflammation ; at the end of this period the eye broke, discharging water (aqueous humor) and matter. After this the pain abated, and the inflammation gradually subsided, till it entirely disappeared by the following October. In the mean time, the patient's sufferings had been so severe as to wear her down very much, affecting her general health, and making her thin, and pale, and weak. Much of the ophthalmic enlargement became permanent ; that is, the original swelling increased very rapidly during the late period of activity. During the following winter she got on tolerably well ; her general health improved, but she was occasionally troubled with paroxysms of pain, neuralgic in character, in the left eyebrow, left half of forehead, and the left cheek. The morbid growth had probably attained such a size as to press considerably on the supra and infra-orbital nerves. In the winter of 1856 and '57, she had fever which lasted six weeks, and left her very weak for a long time. This did not seem to effect the local disease one way or the other. The ophthalmic swelling slowly increased in size, and the neuralgic pains in the left eyebrow, forehead, and cheek, gradually increased in severity until I first saw her (July, 1858). For some time before this, she says she had suffered great distress, the paroxysms of pain being frequent and severe. She also says that any considerable amount of pressure upon the protruding mass, or the handling incident to a physical examination of it, produces, besides the supra and infra-orbital neuralgia (by which I mean pains intermittent, and like electrical shocks), an aching pain in the mass itself, so severe as to deprive her of sleep for the following thirty-six or forty-eight hours. A misstep in walking, whereby the diseased parts are jarred, produces a similar distress, varying only in intensity. She is not subject to any other pains in the head than those mentioned ; except, perhaps, more or less of a sense of distension in the orbit. She has partial deafness in the left ear, but was not aware of it till I ascertained it during the examination. She menstruated at 14, and has always been regular. There is no hereditary disease in the family.

Objective Symptoms.—The patient is pale, and rather thin ; the features are sharpened, and the habitual expression of countenance is that of much distress ; the eye (sound one) is grayish-blue in color, and the hair brown. The patient sits with her head dropping partially forward, apparently from the weight of the tumor.

On close inspection, the protruding mass is seen to be regular in shape, rounded, elongated, and somewhat conoidic at the end, and to hang down almost to the level of the nostril ; the

upper eyelid is thrust forward and stretched down over it to the breadth of two inches; the tarsus and ciliæ are sound; the skin is natural in color; and the eyelid, notwithstanding it is stretched so much, is every where movable, in some degree, over the morbid growth; and near the tarsal border is a vein enlarged, blue, and tortuous. At the opposite, or conoidic end of the tumor is a pearl-colored, nearly circular spot, three fourths to seven eighths of an inch in diameter, evidently the opaque remains of the cornea, which had been destroyed in 1853. Between this pearl-colored, opaque spot, and the tarsal border of the lid, the tumor is dark red in color, and is covered with a granular membrane, which secretes a muco-purulent fluid; this is evidently the ocular and palpebral mucous membrane, pushed down from its natural situation, and stretched over the morbid mass; this dark-red, granular, and thickened conjunctiva is not reflected beneath the eyelid, but terminates abruptly at the tarsal border thereof; no ulcerations are present, and this mucous membrane is, in some degree, movable (non-adherent) throughout its whole extent. Beneath the middle of the upper eyelid, there is a semi-fluctuating sensation imparted to the touch (deep-seated fluctuation). In every other part the tumor has a tense, fleshy, and elastic feel. The orbit is completely filled by it; the orbital margin is well defined, but the size of the orbit itself is considerably increased in the direction of both its perpendicular and horizontal diameters; indeed, the orbit is so much enlarged, that the portions of the malar and temporal bones, which form the external boundary of the orbit, bulge out so much as to produce quite an angular appearance of that side of the head and face. The tumor also seems to be slightly movable in the orbit, but of this I am not entirely sure. I can not discover any disease in the walls of the orbit besides the enlargement above mentioned, which has apparently been produced by the long-continued pressure of the morbid growth.

Diagnosis.—The object of diagnosis is the discrimination of diseases. "The science of diagnosis," says Louis, "is the most important, most useful, and most difficult of all the divisions of medical science. The discrimination of the peculiar nature of each kind of disease, and of its different species, is the source of curative indications. Without an accurate and precise diagnosis, theory is ever at fault and practice often unfaithful."*

The object of diagnosis as applied to the case in hand, is to determine whether the morbid growth we have been describing

* Vide Chomel's Pathology, p. 273.

is cancerous or benign in character ; whether it is developed from the eye itself, destroying that organ by its progressive enlargement, or from the soft parts behind the eye, occasioning ophthalmic protrusion (exophthalmos) and subsequent destruction of the organ, by mechanical pressure : or, again, whether it is developed from the osseous wall of the orbit, or from parts still deeper seated, *e. g.*, the brain ; in fine, whether excision of the tumor be possible, and, if possible, whether such excision would be beneficial to the patient. If the disease is benign, then its extirpation can be attempted with a reasonable expectation of curing the patient : if, on the other hand, it is carcinomatous, no such encouragement should be offered.

The disease resembles carcinoma in several respects. The patient's general health is impaired, and she is thin, and pale, and weak. The swelling also is painful, and located in a situation where most swellings of large size are at once strongly suspected to be cancerous ; indeed, the disease had already been diagnosticated as carcinoma by more than one respectable medical man, and the case pronounced to be utterly hopeless. But the case presented other symptoms, which do not belong to carcinoma, and it was my appreciation of them which led me to a conclusion different from that arrived at by others who had seen the patient. It seemed to me that even the pallor and emaciation might be ascribed to the constant muco-purulent discharge from the conjunctiva, and to the severe neuralgic pain already mentioned, with fully as much propriety as to the constitutional infection of carcinoma ; indeed, the patient's countenance, though pale, did not, as it appeared to me, present that peculiar hue and peculiar expression which are characteristic of the cancerous cachexy. Again : there was nothing about the pain characteristic of malignancy. No pain was occasioned by the tumor, until it became so large as to press upon the ophthalmic branches of the fifth pair that run along the walls of the orbit, and the larger the tumor became, and the greater its pressure upon these nerves, the severer became the pain. The pain did not seem to be in the tumor itself, so much as in the neighboring parts, which were invaded mechanically by the morbid growth. Again : this tumor did not appear to affect the surrounding tissues, otherwise than by pressure. One of the most important characteristics of malignant disease is, as already stated, that it is prone to invade the surrounding tissues by infiltrating them with its own peculiar substance, and imposing upon them its own peculiar structure, thus contracting adhesions to surrounding parts, and, as it were, fusing them all together. The tumor under consideration has done nothing of the kind ; for, notwithstanding its great size, even the skin

covering it is healthy and non-adherent to it. Again: malignant disease about the eyes of long standing, is almost certain to infect the neighboring lymphatic ganglia. In this case, these glands are not involved in any way whatever. And, finally, I believed the tumor to be non-malignant, from the length of time through which its development extended, and the slowness of its growth—from 1851 to 1858, more than 7 years. Objective symptoms showed that, if the disease were malignant, it must be that form known as encephaloid (it was too soft in feel for scirrhus), and encephaloid requiring more than seven years to run its course would be a wonder among surgeons. The statement made by the patient's mother, that no shining, metallic, or yellowish appearance was ever visible behind the pupil, is also of some value in negativing the supposition of ocular cancer.

The tumor was not developed from the eye itself, because that organ lay in front of, and was distinctly movable upon the tumor for a considerable space of time anterior to the attack of conjunctivitis in June, 1853. On this point, both the patient and her mother are positive. The tumor was not developed from the osseous wall of the orbit, because if such had been the case it would have protruded first on the side of the orbit to which it was attached, and it would also have pushed the eye partly in the direction of the opposite side of the orbit; whereas, the tumor actually protruded centrally, and pushed the eye directly before it. The tumor did not involve the brain, because there was no symptom present denoting an organic lesion of that organ. The only cerebral symptom was partial deafness in the left ear, and that entirely lacked proportion to the extent and duration of the disease.

The conclusion appeared to me irresistible, that the protruding mass consisted of a *benign tumor*, which had pushed the eye before it; that said tumor was developed from some of the soft parts behind the eye, and was probably inclosed by the ocular fascia, and mature reflection made these conclusions settled convictions. I did not diagnosticate the tumor as neuroma of the optic nerve, before excising it; for I did not at that time know that neuroma ever attacked the optic nerve, or indeed any other nerve of special sense.

Early in August I expressed to my patient the belief that the disease was not a cancer, but a benign tumor, developed from the soft tissues behind the eye, and circumscribed by the ocular fascia, and that the extirpation of it, though a severe operation and attended with great danger, afforded a reasonable prospect of ultimate recovery. The patient readily assented to my proposal to extirpate it; but as the weather was then hot, and malarious diseases (remittent and bilious-remittent fevers)

were prevalent in her locality (Schuyler's Lake), I advised a postponement of the operation till the weather had become cooler, and the malaria had disappeared.

During the next two and a half months her sufferings increased ; there was more of the feeling of distention in the orbit ; the pain in her left cheek and left forehead came on oftener, was intenser, and lasted longer, producing great distress, and her general health failed considerably.

Operation.—On the 30th day of October, 1858, I proceeded to operate, assisted by Drs. Orendorff, Patrick, Leaning, and Gleason. The patient was placed in the recumbent posture on a couch, the head being raised by means of pillows nearly to the height of an ordinary table. She was then brought fully under the influence of sulphuric ether, and complete anaesthesia was maintained throughout the operation. An incision was made through the skin and orbicularis muscle, from the external canthus, backward and a little upward, to the margin of the orbit. Another incision was made through the thickened mucous membrane, along the tarsal border of the upper eyelid, and close to it across the upper and anterior part of the tumor, from the inner to the outer canthus. The upper eyelid was next dissected off from the tumor, as far as the rim of the orbit, exposing here and there fibres of the orbicularis muscle, and then intrusted to an assistant, to be held out of the way during the subsequent steps of the operation. At the rim of the orbit, the real difficulties of the operation commenced. The cavity of the orbit was so completely filled by the tumor, which, on account of its peculiar attachment, could not be drawn forward, that space could not at first be obtained wherein to detach the tumor in a proper manner. In doing this part of the operation I was desirous of using as little force as possible, for I did not know how much the orbital plate of the frontal bone might have been thinned and weakened, by the long-continued pressure to which it had been subjected, and I was also anxious to do as little injury as possible to the supra and infra-orbital nerves. Indeed, it was not till two cysts of considerable size, containing reddish-brown serum, had been evacuated that the requisite space was attained. (These cysts had occasioned the semi-fluctuation through the upper eyelid heretofore mentioned.) The tumor was then carefully detached from the roof or superior part of the orbit mainly with the handle of the scalpel, dividing here and there a fibrous band with the cutting edge. Then the tumor was drawn upward as far as possible ; the thickened mucous membrane was divided along the tarsal border of the lower lid ; that lid was dissected away, and the tumor was detached from the lower part of the orbit in the

same manner as from the upper part thereof. Its remaining lateral attachments were also cleared. The tumor hanging now only by what seemed to be its root, was drawn forcibly downward and forward, a curved scissors was carried behind it, the root was divided as far back as possible, and then the morbid mass came readily away. A sudden and copious gush of blood from the enlarged ophthalmic artery and vein immediately followed. The hemorrhage was partially checked by the application of cold water, and pressure steadily continued for a few minutes. It was now seen that we had made a clean dissection of the orbit, leaving its cavity, apparently, covered only with periosteum, along which ran certain of the branches of the ophthalmic branch of the fifth pair of nerves, plainly in sight. The lachrymal gland was removed along with the morbid mass above mentioned. The stump formed by the divided ocular muscles being apparently too long, and fearing lest it might afford a nidus for the return of the disease, the curved scissors were again introduced, and it was divided still further back. Then followed a still more copious gush of blood; so great, indeed, as to threaten seriously the patient's life, she having already suffered much from the shock of the operation, together with the previous loss of blood, which had been considerable. Cold, and pressure made with the fingers, appearing to fail in stanching the hemorrhage, the whole orbit was at once carefully filled with compressed sponge, a compress was placed on the outside of it, and a few turns of a roller were wound round the head to secure the whole against displacement. By this means the bleeding was immediately stopped. The patient was readily roused from the anæsthetic sleep by sprinkling her face with cold water.

She rallied slowly from the operation, and complete reaction was not established till late in the following night. But after this no dangerous symptoms supervened. From the risk of inflammation of the brain or its membranes, in addition to the hazards ordinarily attending capital operations, the anti-phlogistic regimen was strictly enforced, and the most absolute quiet was enjoined on the patient. Suppuration was fully established on the fourth day. As no irritation or other difficulty was produced by the sponges, most of them remained in the wound till the sixth day; and the last piece, that is, the one in apposition with the ophthalmic artery and vein, was not removed till the end of two weeks. I hoped by this delay to lesson the risk of secondary hemorrhage. To destroy the fetor occasioned by the sponge remaining in a suppurating wound, a weak solution of liqueur de Labarraque was frequently applied by injection and otherwise. After suppuration was fully estab-

lished, the patient was supported by tonics, of which the most useful was sulphate of quinine and sulphate of iron, and a diet nutritious and easy of digestion. She steadily improved, and in three weeks began to leave her room, and in six weeks to ride. The only difficulty in the after-treatment was occasioned by the sponges, small pieces of which adhered to the deeper parts of the wound, and subsequently, by their presence, produced irritation, exuberant granulations, and retarded the process of cicatrization. As often as these fungus granulations were observed, they were cut down with caustic, and the bit of sponge removed which had occasioned them. These troublesome pieces of sponge were several in number, and varied in size from a pin-head to a pea ; and, what seemed to me very strange, the parts to which they adhered did not seem to possess the power to cast them off by the process of suppuration or ulceration. Sensation was impaired and even destroyed in a part of the left forehead, from unavoidable injury to some filaments of the supra-orbital nerve, but it was gradually restored again as the cure progressed. After a time the partial deafness in the left ear, already mentioned, also entirely disappeared. The bulging of the cheek occasioned by the enlargement of the orbit likewise slowly diminished. At the time of this writing, fourteen months and a half after the operation, she is in good health, the wound is entirely well, and there is no indication whatever of a return of the disease.

The *extirpated tumor-mass*, on a closer examination, is found to be covered externally by a smooth and shining capsule, formed from a condensed cellular tissue, through which certain of the ocular muscles and certain branches of the third pair of nerves are plainly seen. The ocular muscles themselves are elongated, thinned, and flattened, but not otherwise diseased. Within the ocular muscles, and embraced by them on every side, the true capsule of the tumor is found. It is formed by the expanded and thickened sheath of the optic nerve. That nerve enters the posterior end of the tumor near its centre, and, indeed, it seems to form the tumor by expanding suddenly, abruptly, about equally in every direction. The neuroma itself, apart from all extraneous matter, is as large or larger than a goose-egg. On section it appears to consist of a laminate tissue (fibrous), whose color is of a reddish lemon-yellow. The ruptured cysts, heretofore mentioned, are lined by a smooth membrane. In shape, the neuroma is oval, and somewhat elongated. From its distal end, the optic nerve springs out as abruptly as the stem from a water-melon. It then continues of uniform and normal size about one fourth of an inch to the eye, where it terminates. That organ is flaccid ; the lens is absent

(it had probably escaped when the cornea burst, and the humors were discharged in June, 1853), but its coats do not exhibit any evidences of disease other than the results of the inflammation which occurred at that time. The following illustration exhibits at a glance the relation existing between the eye, the neuroma, and the optic nerve on entering and departing from it, the parts being freed from all extraneous matter.

The optic nerve, at the point where it was divided behind the neuroma, is somewhat enlarged, and its tissue is tinged with a reddish and a yellowish hue; not to the same extent, however, as the neuroma. I regret exceedingly, that circumstances prevented a microscopical examination of the tumor.

EXPLANATION OF DIAGRAM.

A. *The Neuroma.*

B. *Remains of the eye.*

Fig. 1. *The optic nerve before entering the neuroma.*

Fig. 2. *The optic nerve between the neuroma and the eye.*

I saw the patient again five years subsequent to the operation. She was in good health, and there had been no return of the disease.

November 1, 1863.

